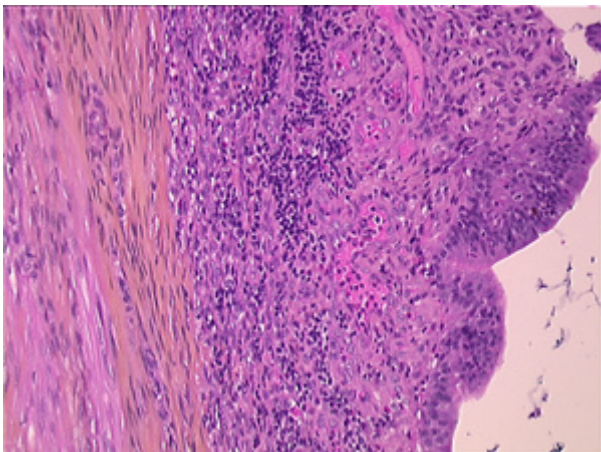
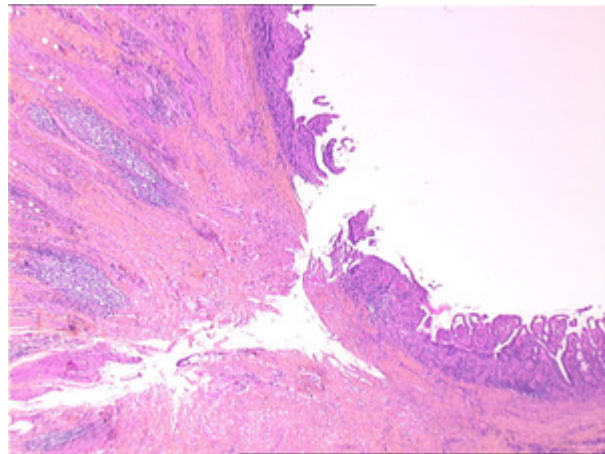
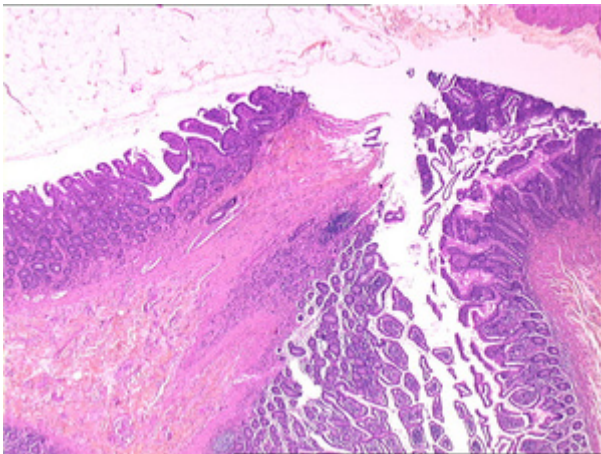


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Iron deficiency anaemia accompanied by obscure abdominal pain for 5 years, in a 57-year-old woman. Capsule endoscopy revealed the presence of two circumferential strictures in the small bowel displaying concentric circular ulcerations and bleeding stigmata. A segmental resection of the small bowel was performed.

What is your diagnosis?



Diagnosis

Diaphragm disease of the small bowel.

Comment

On gross examination, two short circumferential strictures 2 cm distant from each other were observed (Figure 1). These strictures were perpendicular to the long axis of the intestine (Figure 2). On microscopy, in strictures, we observed an exaggeration of the normal plica circularis (Figure 3, first stricture). Mucosa was eroded (Figure 4, second stricture). A chronic inflammatory infiltrate composed of neutrophils and eosinophils was found (Figure 5). Muscularis mucosae was destroyed. Accumulations of hyalinized collagen occupied the submucosa and interdigitated with the muscularis mucosae. This fibrosis contained mild chronic inflammation with small lymphoid follicles. There were no granulomas. No cytomegalic inclusion was found. No lesion of vasculitis was observed.

Multiple mucosal diaphragmatic strictures, also known as mucosal diaphragm disease, was first defined by Lang et al [1] in 1988, who described the pathologic findings of non-specific small-bowel disease in patients taking non-steroidal anti-inflammatory drugs (NSAIDs). Abnormalities of NSAID enteropathy include inflammation, erosion, fibrosis, stricture, perforation, and formation of diaphragm disease. The diaphragm is formed by chronic submucosal fibrosis and destruction of the lamina muscularis due to chronic ulceration, and causes obstruction of the lumen in the small bowel. Diaphragm disease is the most severe stage of NSAID enteropathy.

Diaphragm disease is a rare diagnosis affecting most frequently the ileum. It can also affect the jejunum and the colon, as well as the stomach and the duodenum. The most frequent manifestations are iron-deficient anemia, acute hemorrhage, perforation and obstruction of the small bowel.

The histopathological characteristics of diaphragm disease are: superficial ulceration at the apex of the villi; circumferential ring-like stricture; multiple, short segment annular strictures; transmural inflammation; and submucosal fibrosis. Usually, multiple diaphragms are found in the same time. The depth of ulcer is restricted to the submucosal layer and never extends to the proper muscular layer. The mucosa may show nonspecific inflammation, mucosal eosinophilia and increased numbers of apoptotic cells in the crypt bases. Clinical and/or endoscopic improvement appear after the cessation of the use of NSAIDs, except for diaphragm disease.

In case of multiple small intestinal ulcerations and strictures, the differential diagnosis is cryptogenic multifocal ulcerous stenosing enteritis (CMUSE), whose pathophysiology is unknown. It is characterized as an atypical type of vasculitis. Its clinical features include unexplained stricture and ulceration of the small bowel, affecting only the mucosa and submucosa, without systemic inflammation, and are found in young and middle-aged patients. CMUSE can relapse chronically or after surgery. The symptoms of CMUSE can be improved upon steroid uptake [2]. CMV infection and eosinophilic gastroenteritis can also mimic diaphragm disease of small bowel.

Diaphragm disease of the small bowel, has also been reported in patients with no history of non-steroidal anti-inflammatory drugs intake [3]. Wang et al. recently reported a series of 5 patients (2 men and 3 women) presenting symptoms of gastrointestinal bleeding, characteristics of diaphragm-like strictures of small bowel that were not attributable to the administration of NSAIDs [4]. However, these strictures were slightly different from those induced by NSAIDs with regard to many aspects such as location (middle or distal segment of ileum), number (usually single, no more than three, fibrosis (mild or moderate) and the disease process (non-self-limiting). The cause of these special diaphragm-like strictures remains uncertain.

For further reading

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